

A CONTRIBUTION TO THE PATHOLOGY AND
MORBID ANATOMY OF AMYOTROPHIC LA-
TERAL SCLEROSIS, WITH A REPORT OF
TWO CASES WITH AUTOPSY, IN ONE OF
WHICH, OBSERVED BY DR. C. L. DANA, A
TERMINAL TUBERCULAR CERVICAL MY-
ELITIS AND PERFORATING NECROSIS OF
THE DORSAL CORD WAS FOUND.

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ABSTRACT.

After referring to the moderately constant clinical picture of amyotrophic lateral sclerosis, and the variable pathological conditions, on which they have been founded, the author referred to the fact that the number of cases in the literature, which were well substantiated by autopsy, were fewer in number than one might be inclined to think; a moderately thorough search of the literature revealing only seventy-two cases, which satisfied these conditions. Reference was also made to the different views held by the followers of Charcot and Erb on the one hand, and Gowers, Leyden and others on the other, as to whether the disease is primarily of the pyramids with a secondary involvement of the anterior horns, or whether it is but a variation from the common form of progressive muscular atrophy. The opinion was advanced that, although there was probably more evidence in favor of the former view, this question cannot be settled until a very much larger number of cases, which have been verified by careful histological studies, have been recorded.

The history of two carefully observed cases was then given. The first case was a male, 33 years old. Russian. No pertinent family or personal history. No attributable cause for illness, except overwork and conditions which deprave nutrition, viz. poor and insufficient food, unwholesome and crowded surroundings, and excessive cigarette smoking.

First symptoms in summer of 1893, consisting of

pain and stiffness in neck, headache, vertigo; later stiffness in face muscles, loss of dexterity in fingers of right hand, and later in left; inability to open mouth, dysarthria, dysphagia, nasal twang, increasing difficulty in locomotion on account of stiffness and spasticity.

Examination.—Masked face, considerable atrophy of hands, thenar, hypothenar eminences and interosseous spaces, some atrophy of forearm, more marked on right than left side. Spasticity of upper extremities. Spontaneous and stimulatable fibrillary twitchings. Tendon reflexes all exaggerated. Occipital phenomenon, jaw jerk, triceps reflex all pronounced. Tenderness when atrophied muscles subjected to light squeezing. Cannot walk unless supported. Hyperæsthesia of lower extremities to all forms of stimulation, except faradic. Strong faradic current, such as painful to normal individual, agreeable to him. No real palate pharyngeal anæsthesia, and palate reflex preserved. Special senses normal. No vesical disturbance. Physical condition good. Has peculiar inhibition of emotional reflex, spontaneous and uncontrollable waves of smiling and attempts at laughter. Electrical examination, diminution of faradic irritability, quantitative lessening of galvanic. Unwieldiness of tongue, difficulty in swallowing, respiration weak, pulse rapid. Rather rapid progression of all the symptoms, and after few months the lower extremities began to atrophy. All the symptoms became extremely aggravated, and the patient became bedridden, and death occurred from failing heart and pulmonary œdema.

Autopsy showed no marked deviation from the ordinary in the outward appearances of the spinal cord and brain. The anterior root nerves and hypoglossal nerves were small. On cutting the cervical cord across there was seen a reddish brown discoloration of the anterior horns from the third to the sixth segments, and the consistency of this area was lessened. The entire motor system from cortex to periphery prepared for examination by the methods of Marchi and Algeri, and the Nissl, the Weigert, carmine, nigrosine, hæmatoxylin dyes, etc.

In the spinal cord massive atrophy of the ganglionic cells throughout the cord; best preservation of the cells in the lumbar region, but great diminution of numbers and degeneration of cells here. Degeneration of the crossed and direct pyramidal tracts; few fibres were left undegenerated. Marchi stain shows that the de-

generative process has reached its completion. Throughout the cord, but especially in cervical and dorsal region, evidences of excessive vascularity in shape of large thickened blood-vessels, especially in the gray matter and spaces from which vessels have dropped. In the cervical cord corresponding to the place of apparent softening in the recent state was found great disorganization of the ground substance of the anterior horns.

In the medulla degeneration of the hypoglossal nucleus throughout its entire extent, with the exception of the extreme cephalad end. Slight degeneration in the tenth and common vago-accessorio-glosso-pharyngeal nucleus. No degeneration of the pyramids of the medulla. Root of the 12th nerve small and delicate.

Marchi or Weigert method does not reveal any degeneration in the motor tract from the end of the medulla to the cortex. Nissl's stain shows the cells of the cortex to be normal.

CASE2.—Male, 48, piano maker. Denies alcoholic habits and syphilis, and is of good family and personal history. In October, 1893, first noticed a temporary difficulty and indistinctness of speech and later weakness in left arm; this latter became gradually worse; speech difficulty bettered, but recurred later and remained permanent. Later legs became weaker, and had difficulty in walking. In January, 1894, speech trouble was very slight and arm trouble most prominent. Examination showed that the hand muscles were most affected; the arm and shoulder muscle next. Unable to flex or extend fingers except feebly. Shoulder, arm and forearm muscles all show decided atrophy, but this did not affect any particular physiological groups. No contractures; no fibrillary contractions. Tendon reflexes all greatly exaggerated. No sensory disturbances. Some rigidity of the elbow and shoulder. No disturbances of special senses. Peculiar spasmodic movements of the lips, tongue and jaws; more particularly of tongue; movements as though he were licking lips, or chewing movements—these all beyond his control. Speech disturbance appeared as though due to stiffness and spasm of the tongue. Speech not exactly syllabic. June 15th, 1894—chilliness, fever and weakness; afterward continual elevated temperature, 102 to 104; paralysis in arms and legs increased, little pain in back, slight anæsthesia of legs and arms for short time preceeding death; ten days later unable to speak or swallow.

The spinal cord on section in the fresh state was found to be soft in the neighborhood of the upper dorsal region, and on cutting it down through the dorsal region a spot of white softening could be traced along through the posterior columns. This perforating softened area grew smaller and ended in neighborhood of the lumbar cord. A piece of cord from the cervical region placed in alcohol and culture made from it revealed the presence of the tubercle bacillus in considerable numbers. In cervical region on account of the terminal tubercular myelitis sections, hardened in Müller's fluid and prepared for hæmatoxylin, were very unsatisfactory, but degeneration in the crossed and direct pyramidal tracts with almost complete destruction of the anterior horns could be made out. In the dorsal region there was sclerosis of crossed pyramidal tracts, slight sclerosis of ascending cerebellar tract, diffusely distributed dilated blood-vessels, some with very much thickened walls. Extensive atrophy of cells of anterior horns. In the dorsal region there was found an artificial displacement of the columns of Clarke on one side, so that the column was in juxtaposition with the column of other side. Some degeneration in lumbar and sacral regions as in dorsal, but in lesser degree. In medulla atrophic changes in the nucleus of the 12th nerve. Hæmorrhage of ancient date in dorsal region.

The findings of these two cases were then analyzed and commented on in detail.

The first case would seem to give testimony in favor of the view enunciated by Charcot, insomuch as the Marchi method showed that the degeneration was complete and apparently of somewhat ancient date, whereas the changes in the gray matter were still going on. The intimate and dependent relationship existing between the striking vascular changes and the extensive degenerations, were then discussed, particularly in reference to what may be called the neuron theory. Specimens and drawings of the pathological changes described were exhibited.

Dr. PUTNAM, of Boston.—There is one point of physiological interest which Dr. Collins referred to, on which I should like to say a single word and that is the emotional characteristic which this patient exhibited in connection with these lesions in the medulla. I suppose we have all noticed the extreme emotional tendency of patients with glioma in that region. I certainly have

seen this in several instances, and Oppenheim has recently called attention to the emotional loss of control in patients with bulbar paralysis. I think he referred more especially to pseudo-bulbar paralysis, but at any rate in connection with a disturbance in the action of those parts which are related to emotional display, the emotional sensations, feelings become prominent, and in connection with the recent psychological views advanced by Prof. James and others, I think, that point is one of considerable interest.